CONGENITAL HEART DISEASE

Aberrant tendinous chords with tethering of the tricuspid leaflets: a congenital anomaly causing severe tricuspid regurgitation

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Objective: To define the entity of tricuspid regurgitation caused by tethering of the tricuspid valve leaflets by aberrant tendinous chords.
Design: Retrospective study.
Setting: Tertiary care centre (university teaching hospital).
Patients: 10 patients with unexplained severe tricuspid regurgitation.
Methods: The last 13 500 echocardiographic studies from our facility were reviewed to identify patients with severe unexplained tricuspid regurgitation. Tethering was defined by the presence of aberrant tendinous chords to the tricuspid valve leaflets limiting the mobility of the tricuspid leaflet and resulting in incomplete coaptation and apical displacement of the regurgitant jet origin. Aberrant tendinous chords were defined as those inserting at the clear zone of the tricuspid leaflet and not originating from the papillary muscle. Patients fulfilling the diagnostic criteria for Ebstein's anomaly were excluded.
Results: 10 patients with aberrant tendinous chords tethering one or more tricuspid valve leaflets were identified. There were short non-aberrant tendinous chords in seven patients, five of whom also had right ventricular or tricuspid annulus dilatation.
Conclusions: Tethering of the tricuspid valve leaflets by aberrant tendinous chords can be the sole mechanism of congenital tricuspid regurgitation. It is often associated with short non-aberrant tendinous chords, which may develop secondary to right ventricular or tricuspid annulus dilatation. Awareness of tethering as a cause of tricuspid regurgitation may be important in planning reconstructive surgery.

A degree of tricuspid valve regurgitation is found on Doppler echocardiography in the vast majority of otherwise healthy adults.[1] It can be classified as physiological, primary, or secondary. Primary tricuspid regurgitation results from congenital or acquired morphological anomalies of the tricuspid valve apparatus,[2] while secondary regurgitation is caused by dilatation of the annulus because of right ventricular volume or pressure overload with otherwise normal tricuspid valve morphology.[3] Tricuspid regurgitation may be classified as physiological in the absence of any primary or secondary form of tricuspid regurgitation (fig 1A).

Various congenital tricuspid valve malformations can cause tricuspid regurgitation. These include tricuspid valve prolapse, Ebstein's anomaly, isolated dysplasia of the tricuspid valve, cleft tricuspid leaflet, unguarded tricuspid valve orifice, dysplasia of the left sided tricuspid valve in congenitally corrected transposition of the great arteries (atrioventricular and ventriculo-arterial discordance), atrioventricular septal defect (common atrioventricular junction), or pulmonary atresia with intact ventricular septum.[4] Asymmetrically short tendinous chords to the septal leaflet have also been reported to cause congenital tricuspid regurgitation.[5]

Here we report 10 patients in whom a diagnosis of severe tricuspid regurgitation caused by tethering by aberrant tendinous chords was made by echocardiography. Although tethering of the tricuspid valve has been found in tricuspid regurgitation from other causes,[6 7] we are unaware of any previous reports describing this malformation as the sole mechanism of regurgitation. We will define the diagnosis of tricuspid valve tethering in the context of the normal and pathological anatomy of the tendinous chord apparatus of the tricuspid valve, and distinguish it from other malformations involving tethering.

METHODS

All 13 500 echocardiographic studies performed at our laboratory between January 1999 and March 2002 were reviewed to identify patients with unexplained severe tricuspid regurgitation. Videotape recordings of these examinations were re-evaluated to identify the mechanism of tricuspid regurgitation. This was possible because in all patients with unexplained findings, multiple and partially atypical imaging planes are routinely recorded by the same cardiologist (RJ) in our laboratory. All three tricuspid valve leaflets were thoroughly inspected for the presence of aberrant tendinous chords and short non-aberrant tendinous chords.[5] The normal anatomical distribution of tendinous chords in the tricuspid valve is summarised in table 1.

Tethering was defined by the following anatomical and functional criteria:

- The presence of aberrant tendinous chords to one or more tricuspid valve leaflets (fig 1B). The tricuspid valve leaflets are divided into four zones: the free edge, the rough zone, the clear zone, and the basal zone. The rough zone describes the area between a leaflet's free edge and its line of closure. It is termed ''rough'' because the majority of tendinous chords insert into it. In contrast, the clear zone is thin and translucent, stretching from the line of closure to the basal zone of the leaflets, where the basal chords insert.[8] Aberrant tendinous chords were defined as

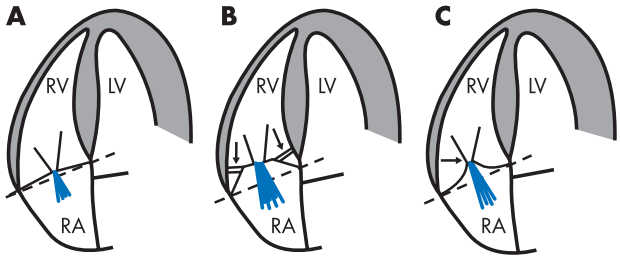

Figure 1 (A) Schematic view of the normal tricuspid valve apparatus with physiological regurgitation; the regurgitant jet originates at the level of the annulus. (B) Aberrant tendinous chords (arrow) tethering the tricuspid leaflets resulting in impaired mobility, incomplete coaptation, and apical displacement of the regurgitant jet origin. (C) Short non-aberrant tendinous chords inserting at the normal site at the tricuspid leaflet free edge, also causing incomplete closure of the tricuspid leaflets with apical displacement of the regurgitant jet origin. LV, left ventricle; RA, right atrium; RV, right ventricle.

inserting at the clear zone of the tricuspid leaflet and originating from the ventricular wall instead of the papillary muscle.

- Incomplete coaptation and impaired mobility of the tricuspid leaflets with tricuspid regurgitation resulting from tethering of the tricuspid valve leaflets.
- Apical displacement of the origin of the regurgitant jet (fig 1A, B).

Short non-aberrant tendinous chords have been defined previously[5] as short tendinous chords inserting at their normal location at the tricuspid leaflet free edge, connecting to the papillary muscle by a tendinous chord that is too short, resulting in incomplete closure of the tricuspid leaflets (fig 1C). In this situation the tricuspid leaflet tips close apically from the annulus.

Table 1 Classification of tendinous chords of the tricuspid valve (adapted from Silver and colleagues[8])

Chord	Site of insertion
Leaflet chords	
1. Rough zone chords	Rough zone and free margin
2. Free edge chords	Free edge
3. Deep chords	Rough zone
4. Basal chords	Basal zone (2 mm perimeter of the annular region)
Interleaflet chords	
1. Fan shaped chords	Commissures between the leaflets

Patients with Ebstein's anomaly were excluded. Ebstein's anomaly was defined by the presence of apical displacement of the septal tricuspid valve leaflet by $\geqslant 8$ mm/m^2 body surface area and of an elongated and redundant anterior leaflet.[7 9]

RESULTS

Among the 13 500 echocardiographic studies analysed, severe tricuspid regurgitation was present in 229 cases (1.7%). Of these, 219 had recognised and defined primary or secondary aetiologies. The 10 patients (five male, five female, mean age 38 years, range 12–73 years) with initially unexplained severe tricuspid regurgitation all had aberrant tendinous chords tethering one or more tricuspid valve leaflets (table 2, figs 2, 3, and 4). Tethering with impaired leaflet mobility resulting in incomplete coaptation was the only mechanism of tricuspid regurgitation in three patients, whereas it was associated with short non-aberrant tendinous chords in seven (table 2). Among these 10 patients, secondary dilatation of the right ventricle (apical four chamber view) was present in four, and of the tricuspid annulus in seven. Five of the seven patients with short non-aberrant chords in addition to tethering had either right

Table 2 Morphological characteristics of patients with severe tricuspid regurgitation caused by tethering

Patient number	Sex	Age (years)	Other congenital cardiac abnormalities	ΔP syst RV−RA (mm Hg)	Tricuspid annulus diameter (mm, normal <31 mm)	Area-D (cm^2)	Fac (%)	Origin of the TR jet displaced	Leaflets with tethering from aberrant chords			Leaflets with short primary chords		
									Ant	Sept	Post	Ant	Sept	Post
1	F	12	–	25	45	32.0	42	Apical	–	+	+	–	–	–
2	F	56	–	28	25	18.7	49	Apical	–	+	–	–	+	–
3	F	73	–	39	35	14.0	46	Apical	+	+	–	–	–	–
4	M	18	–	21	44	24.0	20	Apical	–	+	+	–	+	+
5	M	17	Superior sinus venosus septal defect	15	39	20.1	48	Apical	+	+	+	+	+	+
6	M	41	AV/VA discordance (*l*-TGA)		21	27.1	12	Apical	–	+	+	–	+	+
7	M	35	AV concordance/ VA discordance (*d*-TGA)		52	40.9	48	Apical	–	+	–	–	–	–
8	M	62	Situs inversus with mesocardia; AV/VA discordance (*d*-TGA)		48	33.0	69	Apical	–	+	–	–	+	–
9	F	43	Pulmonary stenosis, ASD II	45	35	14.2	56	Apical	+	+	+	+	+	+
10	F	20	Pulmonary stenosis	30	30	13.5	44	Apical	–	+	+	+	+	+

Ant, anterior; Area-D, end diastolic area of the right ventricle obtained in the apical four chamber view (normal <25 cm^2); ASD, atrial septal defect; AV/VA discordance, atrioventricular and ventriculo-arterial discordance; AV, atrioventricular; Fac, fractional area change (normal >25 cm^2) defined as end diastolic area minus end systolic area divided by end diastolic area of the right ventricle; Post, posterior; RA, right atrial; RV, right ventricular; Sept, septal; syst, systolic; TGA, transposition of the great arteries; TR, tricuspid regurgitation; ΔP, difference in pressure; VA, ventriculo-arterial.

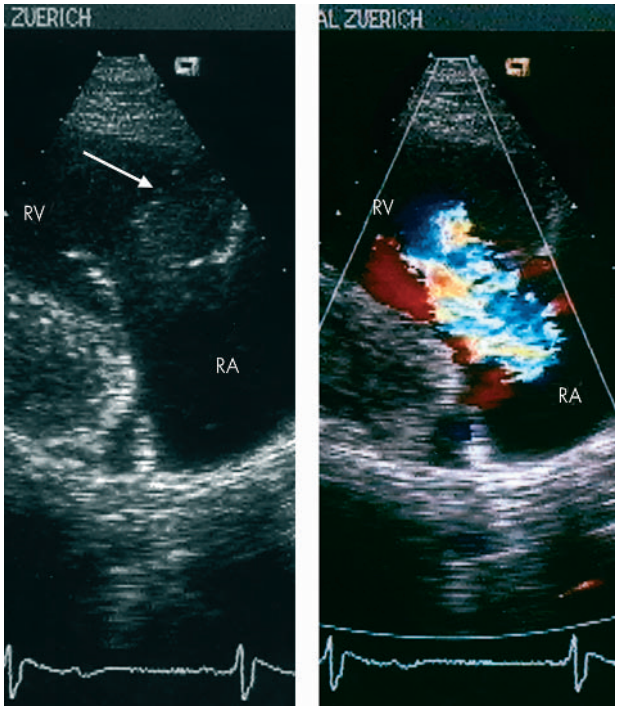

Figure 2 Echocardiographic atypical parasternal long axis view of the tricuspid valve apparatus. Left panel: Arrow indicates an aberrant tendinous chord tethering the anterior tricuspid leaflet. Right panel: Severe tricuspid regurgitation is detected by colour Doppler imaging, and apical displacement of the regurgitant jet origin is apparent. RA, right atrium; RV, right ventricle.

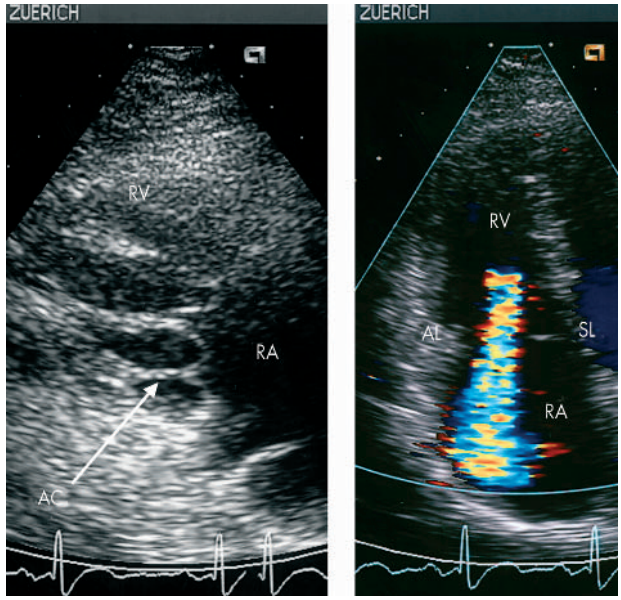

Figure 4 Left panel: Echocardiographic atypical parasternal long axis view of the tricuspid valve apparatus showing an aberrant chord (AC) tethering the posterior tricuspid leaflet. Right panel: Severe tricuspid regurgitation is detected in the same patient by colour Doppler in the apical four chamber view; the origin of the regurgitant jet is displaced towards the apex. AL, anterior tricuspid leaflet; AL and SL, tricuspid annulus plane; RA, right atrium; RV, right ventricle; SL, septal tricuspid leaflet.

ventricular or tricuspid annulus dilatation, or both, while two had neither. Interestingly, short non-aberrant chords were not found in the patient with the largest right ventricle and tricuspid annulus (patient 7).

Tethering was associated with a congenital cardiac abnormality in six patients (table 2): one patient had a superior sinus venosus atrial defect; one had atrioventricular and ventriculo-arterial discordance (congenitally corrected *l*-transposition of the great arteries); one had atrioventricular

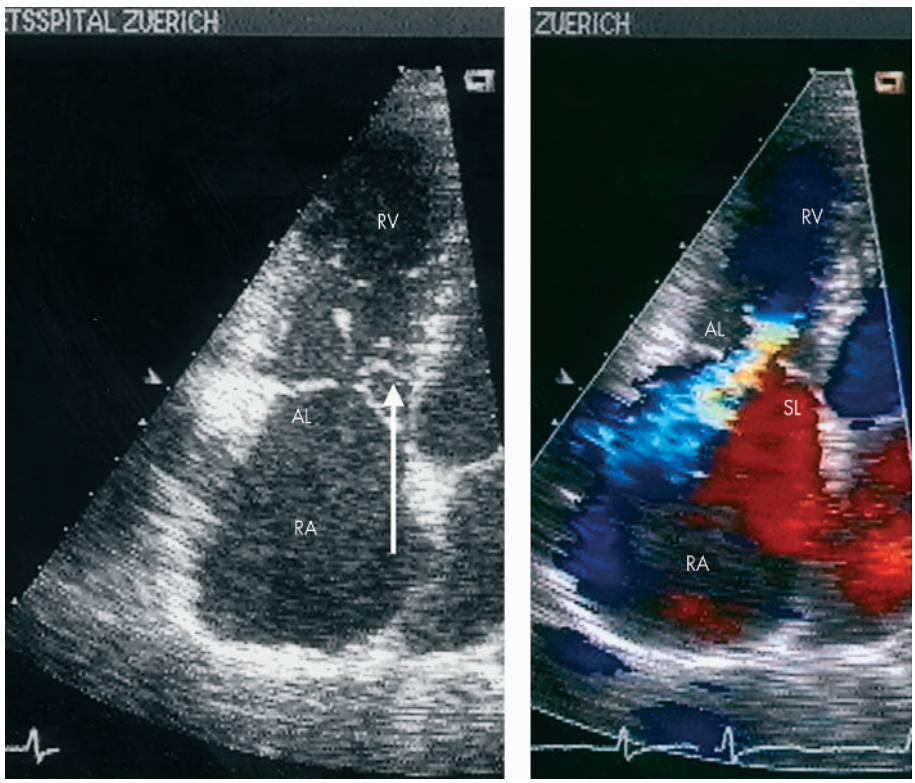

Figure 3 Apical four chamber view of the tricuspid valve apparatus. Left panel: Arrow indicates an aberrant tendinous chord originating from the ventricular septum and tethering the septal tricuspid leaflet. Right panel: Severe tricuspid regurgitation is detected by colour Doppler imaging. Note the apical displacement of the regurgitant jet origin from the annulus plane. AL, anterior tricuspid leaflet; AL and SL, tricuspid annulus plane; RA, right atrium; RV, right ventricle; SL, septal tricuspid leaflet.

concordance and ventriculo-arterial discordance (*d*-transposition of the great arteries) and underwent a Blalock-Hanlon atrioseptectomy at four days and a Mustard procedure at eight years; one had a situs inversus with mesocardia and atrioventricular and ventriculo-arterial discordance (with *d*-transposition of the great arteries); and two patients had pulmonary valvar stenosis and had undergone commissurotomy at the ages of 5 months (patient 10) and 4 years (patient 9).

DISCUSSION

The normal anatomy of the tricuspid valve chord apparatus is complex and has been described in detail previously (summarised in table 1).[8 10–12] Chords are classified by their morphology and site of insertion into the tricuspid leaflet. We have defined tethering anatomically and functionally by the presence of aberrant tendinous chords, which results in impaired mobility, incomplete coaptation, and apical displacement of the tricuspid leaflet tips during systole (fig 1B). As a result, the origin of the regurgitant jet is displaced towards the apex compared with other mechanisms of tricuspid regurgitation. Aberrant tendinous chords insert at the clear zone of the tricuspid leaflet—which is usually free of chord insertions—instead of the usual site of insertion at the free edge or rough zone, and connect to the endocardium instead of the papillary muscle. They are distinct from basal chords, which also arise from the endocardium, but insert into the leaflet only in close proximity to the annular region (maximum 2 mm).[8] One or more of all three tricuspid valve leaflets may be affected by tethering. In fig 5 normal and aberrant tendinous chords are shown in pathology specimens.

Tethering of the tricuspid valve is a prominent feature of Ebstein's anomaly. Among 41 patients with this disorder described in detail by Shiina and colleagues,[6] 35 had tethering of the septal and anterior tricuspid leaflets. However, diagnostic criteria for Ebstein's anomaly set up by the same group rely on the apical displacement of the septal leaflet by at least 8 mm/m^2 and the presence of an elongated anterior leaflet.[7] Tethering was not included in the diagnostic criteria, and the presence of tricuspid valve tethering has been a frequent source of mistaken diagnosis of Ebstein's anomaly.[7] Tethering can also be distinguished from short, non-aberrant tendinous chords as a cause of tricuspid regurgitation.[5] Short non-aberrant chords connect the papillary muscle to the leaflet free edge, but are too short, resulting in incomplete closure of the tricuspid leaflets (fig 1C) and apical displacement of the leaflet tips from the annulus plane during systole. Thus, this pathology results in a functional defect similar to tethering, but by chords in an anatomically correct location. This architectural difference has implications for surgical repair; while aberrant tethering chords may simply be severed, thus restoring normal leaflet coaptation, resection of short non-aberrant chords would result in a flail tricuspid leaflet. Aberrant tendinous chords with tethering may occur alone or in combination with short tendinous chords. These two malformations were found to be associated in 70% of our cases. Both entities may affect each of the three tricuspid leaflets. However, distinguishing short chords as a primary congenital defect leading to tricuspid regurgitation from chords which have become too short in the course of right ventricular or tricuspid annulus dilatation from other causes may be difficult. This was apparent in our series, in which five of seven patients with short chords had associated dilatation of the right ventricle or tricuspid annulus. Thus in some cases a possible sequence of events might be the development of right ventricular or tricuspid annulus dilatation caused by severe tricuspid regurgitation from tethering as a primary defect, which then leads to short non-aberrant chords.

Many congenital tricuspid valve anomalies can cause tricuspid regurgitation. Independent of classification, which in addition may share some overlap owing to imprecise diagnostic criteria, awareness of tethering as a mechanism of tricuspid regurgitation is of clinical relevance and should be considered in cases of regurgitation with impaired leaflet mobility and apical displacement of the regurgitant jet origin. An updated classification of tricuspid malformations causing regurgitation is proposed in table 3.

Surgical repair of a regurgitant tricuspid valve is carried out according to the principles established for mitral valve repair.[13 14] Reduction of the valvar annulus is often the only necessary measure to correct the great majority of secondary tricuspid valve insufficiency. In patients with tethering by aberrant chords, however, the reduction of the annulus alone may not reduce the regurgitation sufficiently. Although to date we have no experience of this, we believe that resection of the tethering chords could restore full mobility to the

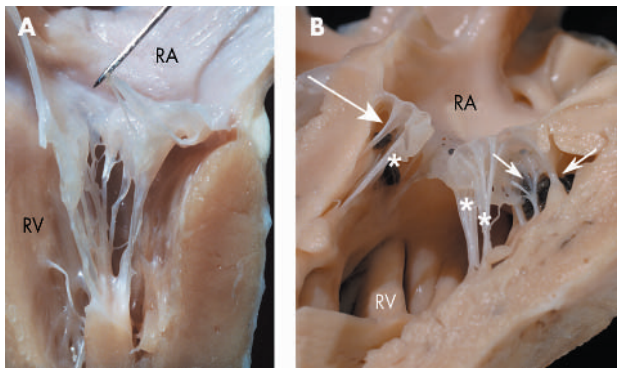

Figure 5 (A) Pathology specimen with view of the posterior tricuspid valve apparatus is shown in a normal heart without aberrant chords; the tendinous chords insert normally at the papillary muscle. (B) Pathology specimen with an aberrant tendinous chord (long arrow) tethering the septal tricuspid leaflet. Normal basal chords (short arrows) are visible inserting at the anterior leaflet. Normal rough zone chords are marked with an asterisk. RA, right atrium; RV, right ventricle.

Table 3 Aetiology of tricuspid regurgitation

1. Primary tricuspid regurgitation	
(A) Congenital	Tricuspid valve prolapse
	Ebstein's anomaly
	Tricuspid valve dysplasia (grades I–III)
	Abnormal number of leaflets
	Atrioventricular channel
	Cleft of a tricuspid leaflet
	Left sided tricuspid valve in congenitally corrected transposition of the great arteries
	Pulmonary atresia with intact ventricular septum
	Short tendinous chords
	Aberrant tendinous chords with tethering of the tricuspid leaflets
(B) Acquired	Endocarditis
	Rheumatic valve disease
	Right ventricular infarction
	Heart transplantation
	Carcinoid (or other tumours)
	Trauma
	Rheumatic arthritis
	Radiation therapy
	Papillary muscle dysfunction
	Hypereosinophilic syndrome
	Thyrotoxicosis
	Anorectic drugs
2. Secondary tricuspid regurgitation	
3. Physiological tricuspid regurgitation	

involved leaflet, which could then reach the coaptation plane. Although it may be possible in some cases to remove the tethering chords by pulling them out between primary chords with a nerve hook, the best way to access them may be by detaching the involved leaflet from the annulus, resecting the restricting chords, and then reinserting the leaflet to the annulus. Recognition of this entity and a precise definition of the tricuspid lesion are therefore essential to tailor a specific and appropriate surgical repair.

CONCLUSIONS

Tethering of tricuspid valve leaflets by aberrant tendinous chords may, in rare cases, be the sole mechanism of severe tricuspid regurgitation. Awareness of this mechanism is of clinical relevance and should be taken into consideration when planning reconstructive surgery of the tricuspid valve for tricuspid regurgitation.

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FROM BMJ JOURNALS

Myocardial perfusion scintigraphy and coronary disease risk factors in systemic lupus erythematosus

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Objective: To evaluate the prevalence of myocardial perfusion abnormalities and the possible association between myocardial perfusion defects and traditional coronary artery disease (CAD) risk factors as well as systemic lupus erythematosus (SLE) related risk factors.
Patients and methods: Female patients with SLE, disease duration >5 years, age 18–55 years, who had used steroids for at least one year were enrolled. Traditional CAD risk factors evaluated were arterial hypertension, diabetes mellitus, dyslipidaemia, postmenopausal status, smoking, obesity, and premature family CAD profile. Myocardial perfusion scintigraphy was evaluated by single photon emission computed tomography with technetium 99m-sestamibi at rest and after dipyridamole induced stress.
Results: Eighty two female patients with SLE without angina pectoris with mean (SD) age 37 (10) years, disease duration 127 (57) months, SLE Disease Activity Index (SLEDAI) score 6 (5), and SLICC/ACR-DI score 2 (2) were evaluated. Myocardial perfusion abnormalities were found in 23 patients (28%). The mean (SD) number of CAD risk factors was 2.2 (1.6). There was a significant positive correlation between age and number of CAD risk factors. Lower high density lipoprotein (HDL) cholesterol level showed a significant association with abnormal scintigraphy. Logistic regression analysis showed that lower HDL cholesterol level and diabetes mellitus were associated with myocardial perfusion abnormalities. Current vasculitis was also associated with abnormal scintigraphy.
Conclusions: Lower HDL cholesterol level and diabetes mellitus have a significant influence on abnormal myocardial perfusion results found in asymptomatic patients with SLE. Current vasculitis was associated with abnormal myocardial scintigraphy. These data suggest that abnormal myocardial scintigraphy may be related to subclinical atherosclerosis.

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